REVIEW ARTICLE

Comprehensive Management of Pediatric Cataract in Africa

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ABSTRACT

Worldwide, childhood blindness is 0.75/1000 children giving an estimated number of 1.4 million suffering from blindness worldwide. Of the blind children worldwide, congenital cataract is the major cause in African countries. The management of cataract in children poses a challenge to the African ophthalmologist, and the earlier in life cataract develops, the more difficult it is to manage. A description of the common causes, clinical presentation, methods of modern surgical removal, and suggested typical postoperative management of this condition is given along with common complications and challenges that may be encountered.

Keywords: Africa, cataract, lens aspiration, Nigeria, pediatric

INTRODUCTION

Worldwide, childhood blindness is 0.75/1000 children giving an estimated number of 1.4 million suffering from blindness worldwide.[1,2] Of the blind children worldwide, congenital cataract is the major cause in African countries and other middle to low-income countries.[3] Thus, Africa has a very high prevalence of blindness of up to 10 times that of industrialized nations.[4]

The management of cataract in children particularly in the visually immature ones poses many challenges to the ophthalmologist, and the earlier in life cataract develops, the more difficult it is to manage. However, the need for early intervention in these cases is well established to prevent visual deprivation amblyopia, especially in infants with uniocular cataracts where it is more profound.[5]

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The introduction and constant improvement of new microsurgical techniques, better and more precise instrumentation, and the introduction of high molecular weight viscoelastics have enabled surgeons to remove cataracts safely at an early age and have improved the outcome significantly. It is therefore important to adopt these new techniques in developing African countries to improve the visual outcome despite the fact that it requires significant financial investment in human resources and relatively expensive equipment in childhood eye care. The political will to do this currently may not be equally available in many African countries.[6]

PATHOGENESIS OF CATARACTS

There is a wide variety of etiologies and appearances of cataracts in children. It may also be unilateral or bilateral. It may be present at birth or appear later in life.

Unilateral infantile cataracts may be sporadic whereas bilateral infantile may have other associated disorders

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and systemic diseases. One-third of cases are idiopathic, however with no apparent cause.

In older children, the cause could be related to trauma, electric shock, or the use of drugs, for example, steroids. Heritable cataracts may be autosomal dominant, autosomal recessive, and/or X-linked.

Over 350 heritable conditions related to cataracts are listed in the Online Inheritance In Man (OMIM) database and can be looked up online. It is not however known how many of the cataracts seen in African countries are heritable.

Medical conditions such as diabetes or galactosemia may be associated with bilateral cataracts.

Infants with bilateral cataracts require full systemic evaluation in addition to genetic evaluation if available. Genetic tests may however not be required when there are only uniocular cataracts.

In case there is a systemic association, all children should be referred to a pediatrician for a full examination.

If ectopia lentis is seen in one eye only, rule out a prior history of trauma. Bilateral ectopia lentis generally has a systemic association which may be life-threatening, which needs to be looked for and addressed by the appropriate specialist.

Systemic conditions associated with cataracts

- Chromosomal abnormalities, for example, trisomy 18 (Edwards syndrome)[7] and trisomy 21[8]
- Infectious diseases, for example, toxoplasmosis, rubella, cytomegalovirus, herpes simplex, and syphilis (TORCHES)[9]
- Metabolic disorders, for example, galactosemia, diabetes mellitus[10]
- Renal disorders - Lowe's and Alport's syndrome[11,12]
- Parathyroid disorders - hyperparathyroidism and pseudohypoparathyroidism in some cases[13]
- Musculoskeletal disorders, for example, in some cases with Myotonic dystrophy[14]
- Others, for example, Cockayne's syndrome,[15] incontinentia pigmenti.[16]

Evaluation/investigation of children with cataracts

The parents should be asked about a family history of cataracts (white spots in the eye). This will help differentiate whether the cataract is congenital, developmental, or traumatic in origin. In addition, ascertain history of exposure of the mother to drugs, rashes, or infection during pregnancy.

A general physical evaluation should be carried out by the pediatrician to rule out systemic associations, anomalies, congenital rubella, and possible fitness for surgery.

It is important to examine siblings and parents especially the mother. She may also have cataracts on slit lamp examination if it is familial. Do not forget to do this. One may find a visually insignificant cataract. For example, the mother (female carrier) of a male child with Lowe's syndrome (which is an X-linked condition) may have punctate lens opacities.

A complete ophthalmic workup in addition to a dilated (portable) slit lamp evaluation of the cataract should also be carried out in every child. There are several relatively cheap models available. Every pediatric ophthalmologist should ideally have one. However if only the adult slit lamp is available, the infant can be carried to place his chin on the chin rest with the rest of its body supported and straightened out in the so-called "flying baby" position.[17,18]

Ocular examination should include visual acuity assessment, pupillary response, and ocular motility. Check for cloudy or enlarged cornea in case of associated conditions such as a coexisting glaucoma or peters anomaly. Biomicroscopy should be carried out in each case to evaluate size, density, and location of the cataract to plan the surgical procedure. Fundus examination should be carried out after pupillary dilatation.

These include the following:

- B-scan ultrasound especially if the posterior segment is not visible. This will rule out posterior segment masses such as retinoblastoma, retinal detachment (RD), persistent fetal vasculature, and a suspected retained intraocular foreign body (IOFB) in a traumatic cataract. If IOFB is still not seen and it is still suspected, a computerized tomogram scan can be ordered.[19]
- Chromosome analysis[20]
- TORCHES titer
- Urine reducing substances-galactose 1 phosphate uridyl transferase, galactokinase
- Urine amino acids
- Serum calcium, phosphorus, and glucose
- Serum ferritin-hyperferritinemia.

These tests will help rule out TORCHES and inborn errors of metabolism.[9]

Evaluation of children with ectopia lentis

Marfan's syndrome

The lens dislocates superiorly even though the zonules are intact and unbroken. Cardiac ultrasound can pick

up coarctation of the aorta which is an important association and potentially life-threatening. Fibrillin gene mutation screening can also be carried out to confirm the diagnosis. RD may occur years later after cataract surgery.

Homocystinuria

The lens usually dislocates into the anterior chamber while the zonules are completely broken. This condition causes thromboembolic phenomena which is a hazard when undergoing anesthesia. Urine screening for disulfides will help clinch the diagnosis.

Sulfite oxidase deficiency

Measuring sulfite oxidase activity in skin fibroblasts confirms the diagnosis in which case activity is usually absent. This condition leads to severe neurological problems which may result in irreversible brain damage and death by age 5 years.

Clinical presentation of pediatric cataracts

1. Cataracts – This causes an abnormality of the red reflex. They are usually described by parents as a "white spot in the eye." The more posterior and central the cataract is, the greater the effect on vision. Those present at birth are the most serious because the visual system is not fully developed, and vision could be lost irreversibly if the visual axis (VA) is not cleared by 6–8 weeks of life[10]
   Bruckner test will usually be positive when a significant cataract is present.[21] This should be taught to all medical/health personnel involved in the care of children. This is usually carried out by shining the beam of a direct ophthalmoscope on both eyes of a patient suspected to have leukocoria and viewing the reflection of the beam back from the retina through the pupils from 40 cm away through the observer piece. Unequal intensity of the red reflex seen through the pupils may mean the child could have cataracts and should be referred to an ophthalmologist or pediatric ophthalmologist if available. This will promote early intervention which in turn will improve visual outcome
2. Poor vision - The mother complains of the child (depending on the age) not smiling back at her or the child bumping into objects
3. Nystagmus usually develops within 2–4 months of life. This is usually seen when visually significant cataracts occluding more than 3 mm of the VA are neglected.[22] It is a poor prognostic sign in terms of visual function
4. Uniocular cataracts develop profound amblyopia or lazy eye due to obscuration of the visual axis
5. Strabismus.

Cataracts can affect any part/layer of the lens.

Assessment of vision in children

This is very important. One needs to know how to handle children to successfully and accurately check vision in them.

It is important to employ age- and child-specific tests.

Children are smart: We just have to be smarter.

One may use direct and indirect methods of assessing vision in them.

Indirect methods

1. Blink reflex in response to sound
2. Menace reflex-closure of eyes when object is approaching if visual acuity (VA) is normal
3. Fixation patterns - check if they are central, steady, and maintained. If they are not, they may be referred to as uncentral, unsteady, and unmaintained. One can check if the child can fix and follow or not
4. Check the child's monocular behavior/is there avoidance of occlusion?
5. Check if he/she makes visually directed movement.

Direct measurement of children with cataracts

In infants or patients with other disabilities:

1. Optokinetic drum
2. Preferential looking (primarily the basis of the Lea grating paddles tests Teller acuity cards)
3. Visual evoked response.

1–2 years:

- Hundreds and thousands (Similar to tiny multicolored cake decorations)
- Marble game test.

2–3 years:

- Miniature toy test
- Sheridan's ball test
- Cardiff acuity test.

Once the child is old enough to read, adult methods (Snellen charts, ETDRS charts, etc.) can be used. However, children are currently being taught phonetic sounds to pronounce letters. Hence, do not be surprised when they start making sounds when reading rather than read out the simple letter K or P, the way it was traditionally read out.

Differential diagnosis based on morphology

Anterior polar cataract

It is usually not progressive but can cause asymmetric refractive errors which lead to amblyopia.[10]

Nuclear cataract

This type is noted in the center of the center of the lens and it usually occurs lens formation.

It is inherited as autosomal dominant. The effect on vision depends on the density and size. It may have a significant effect on vision if it is wider than 3 mm.

Lamellar cataract

This type affects one or more layers of the mid-periphery of the lens.

It is generally larger than a nuclear cataract and develops at a later stage of lens formation than nuclear cataracts.

Posterior subcapsular cataract

These are irregular white opacities just under the final layer of the lens.

They may be present at birth or may be acquired, for example, traumatic, steroid induced, or radiation induced.

Posterior lenticonus

There is usually a weakness of the posterior capsule (PC).

It is usually not present at birth but observed later in childhood, thus it has a better prognosis.

Persistent fetal vasculature

This results from incomplete regression of the hyaloid artery. There is dense white vascularized plaque on PC. The vessels may contract. The ciliary processes are usually seen.

There is usually associated microphthalmia. If the stalk contracts, it may lead to retinal detachment.

In these patients, there is a high risk of glaucoma.

The best visual results are obtained if there is no retinal involvement.

Early treatment of the condition in addition to aggressive amblyopia therapy usually gives the best results.

Other types of cataracts

These cataracts include oil droplet cataract, cataract from congenital rubella syndrome (CRS), anterior lenticonus, Alport's syndrome, Marfan's syndrome leading to subluxated lens, lens in vitreous as seen in homocystinuria, etc.

Treatment

Treatment depends on the age of the child and the degree to which the opacity affects vision. Visually significant cataracts produce a blurred image on the retina which affects the development of the visual pathways.

A good outcome depends on the following:
1. Successful surgical removal of the cataract
2. Replacement of the lens focusing power
3. Treatment of amblyopia.

Uniocular cataracts are notorious for being highly amblyogenic. Therefore, surgery is the best option within 4–6 weeks of life.[23] Surgery before this (<1 month of life) may lead to secondary glaucoma due to the poorly formed anterior chamber angle.[24]

Bilateral cataracts may be amblyogenic-time frame for optimal removal is 2–3 months before nystagmus develops.[10]

Different scenarios of cataracts seen

- Uniocular dense seen in infancy
- Older child with bilateral cataracts since infancy
- Older child with uniocular cataracts since infancy
- Older child with recent development of bilateral cataract [Figure 1]
- Bilateral immature cataracts.[25]

SURGERY

Surgery is indicated when the potential benefits of surgery outweigh the potential risks. The potential risk includes the significant loss of focusing power or ability to accommodate. The aim of surgery is to provide long-term clear axis by preventing the development of Visual axis opacification (VAO).

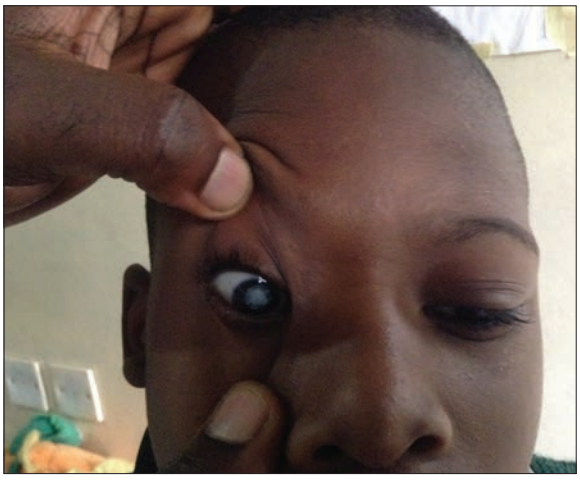

Figure 1: Older child with recent development of bilateral cataract

Objective assessment of the degree to which it affects vision may be done with the (portable) slit lamp.

Evaluation of the red reflex is carried out using either the direct, indirect ophthalmoscope, or a retinoscope.

Poor visualization of the retina indicates the need for surgery.

Assess the posterior segment carefully.

In older children: If VA is 20/40 or worse, consider surgery weighing the risks depending on the instrumentation available.

Prognosis depends on whether the rest of the eye is normal.

Patient preparation

Ideally it is better to admit a day before to observe as surgery is under general anesthesia (GA).

It is important to dilate pupils with 1% tropicamide and phenylephrine or cyclopentolate drops. Rule out associated prior history of convulsions in these patients before using these drops as they can induce one.

Ensure that there are no skin infections, ocular, or ear discharge in the child also rule out coexisting nasolacrimal duct obstruction. These should be treated before surgery.

We have documented physician or pediatric clearance for GA to minimize anesthetic and systemic complications.

The child must be stable systemically in case there are cardiac features, for example, CRSs which may have coexisting heart disease.

Examination under anesthesia

After induction, the child's eyes are examined.

General inspection should be done through dilated pupils.

The following tests should be done:
Keratometry: The shape of the cornea is measured in 2 meridia.

A-scan: This measures the axial length for calculating the intraocular lens (IOL) power and establishing a baseline for monitoring the globe elongation postoperatively.

Horizontal corneal diameter: No IOL should be inserted if cornea is < 10.5 mm diameter.

IOP should be carried out with applanation tonometer. Avoid using fluorescein as it tends to stain the cornea. Rather use povidone iodine as your dye to see the menisci.

Biometry with SRK II formula is preferred according to various studies in children.[26] However, a recent report by the Infant Aphakia Treatment Study (IATS) team suggests that using either the Holladay 1 or SRK/T formula is best.[27] However, the absolute prediction errors have been found to be substantial only in eyes that were <18 mm in axial length regardless of whichever formulae were used.[27]

To have a baseline in case glaucoma develops later, anterior chamber angle assessment should be done with direct gonio lens and recorded.

Problems associated with pediatric cataract surgery

Small eyes

The eyes of a child are 40–50% smaller than that of an adult.[10,28] The mean axial length of a newborn's eye is 17 mm compared to 23–24 mm in an adult.[10,28] The eyes rapidly grow till the age of 11 years with resultant rapid change in refraction making IOL calculation and eventual refraction difficult to predict. Studies have shown that the refractive error of aphakic children undergoes a myopic shift of 7–8D from age 1 to 10 years.[29] The tendency therefore is to undercorrect to cope with the myopic shift. Therefore, lens implantation requires a compromise that accounts for the age of the child and the target refraction at the time of surgery. Most surgeons implant IOLs with powers that will be required in adulthood, allowing the child to grow into the power selection of the lens. Thus, the child is undercorrected and will require hyperopic spectacles of decreasing strength until the teenage years. Making the child emmetropic at time of surgery which some surgeons do for uniocular cataracts (to facilitate the development of binocular function around the time of surgery) will only make the child anisometropic due to progressive myopia that develops over time in these children. It may be best to avoid this.

The use of adult-sized IOLs (which is what may be available in developing countries) is associated with sizing issues particularly in small eyes in the first 2 years of life. The smallest optic (5.5 mm) should be used.

Necessity and risk of GA: This necessitates bilateral simultaneous cataract surgery in some developing

countries.[30] This is however not recommended and should not be a routine.

High rates of postoperative inflammation:
This may in turn cause a secondary glaucoma.

High rates of Visual axis opacification (VAO) have been linked to a high rate of reoperations.[31]

Noncooperation for yttrium aluminum argon garnet (YAG) laser: This is an important reason why an adequate posterior capsulorhexis and anterior vitrectomy (AV) should be carried out initially. However, this has been associated with higher rates of RDs later in life in older studies due to uncontrolled disturbance of the vitreous.[32] More recent studies however have not found AV significantly predictive of RD.[33] Therefore, if at time of surgery, it is observed that in the next 18 months, the child will be able to cooperate for yttrium aluminum garnet (YAG); it is recommended that the PC rather be left intact to obviate this. To cooperate for neodymium-doped: YAG laser capsulotomy, the child should be at least 4 years old.[10]

If funds are limited, one may rather choose to invest in a phaco machine than in, for instance, a YAG laser.

Cost of surgery is high due to the special equipment needed. A phaco machine with capability is needed to aspirate and perform AV. This may be out of the reach, and excellent training may not be available. However without it, the outcome of surgery for children may not be as desired and so every effort should be made to acquire one. However, in the absence of it, skillful small incision cataract surgery could be performed with aspiration of the soft nucleus with Simcoe/Blumenthal cannula. The use of relatively cheaper equipment such as a portable vitrectomy will improve the outcome. It is essential to avoid performing manual sponge vitrectomy.

Frequent change of glasses: This is because the refraction changes as the child grow. The child also may lose them. The child should have refraction at least every 6 months. Plastic well-fitting frames big enough to cover up to the brows with elastic support round the head without nose pads are best for children.

Frequent follow-up: This is important to pick up early signs of problems and to ensure compliance by care givers. They therefore are seen more frequently in the clinics than adults postoperatively.

The surgery itself

The three parts of the lens are handled separately: Anterior lens capsule, lens cortex, and posterior capsule and are discussed in detail in the following sections.

- The anterior lens capsule:[34] Continuous curvilinear capsulorhexis (CCC). This should be made at least 4.5–5 mm wide. The capsular dye, 0.1% trypan blue dye injected into the anterior chamber under air cover facilitates this particularly in white cataracts. It may be better to use a cohesive viscoelastic such as Healon GV to make the CCC easier as it helps maintain the anterior chamber stability, helps offset the low sclera rigidity, and increased vitreous upthrust found in pediatric eyes. Runaway "rhexis" is more common in children due to the highly elastic capsule. Once the [rhexis] is completed, the edges resist [easy] tearing. Avoid using the adult can opener technique. It is best to use an intraocular forceps after making a nick with a capsulotomy needle seen in Figure 2. A "push and pull" technique may be easier in children[10]
- Lens cortex:[34] Lens aspiration complete removal is important particularly in the equatorial region. In some cases, hydrodissection using a 27–30 G cannula under the rhexis margin may facilitate this. However, this should be avoided with posterior lenticonus and in posterior polar cataract. Most of the pediatric cataract can be aspirated using the two-way irrigation aspiration (IA) cannula or a simcoe (if your opening is large enough to admit it) or automated IA on the phaco machine. If the capsule is calcified or membranous, using an intraocular scissors and gently removing it with the intraocular forceps may be better than using phacoemulsification or vitrectomy the vitrector may become easily blunt and they are expensive to replace. Using two ports, one for irrigation and the other for aspiration works very nicely while maintaining the anterior continuous during the procedure. You may use the irrigation probe to

Figure 2: Intraocular scissors - curved and straight

Figure 3: Intraocular forceps - curved and straight

push in the cortex into or unclog the aspiration probe by rubbing the tips together

- Posterior capsule:[34] Primary posterior CCC (PPCCC) should be made at most 3.5–4 mm wide as VAO is the most common complication after a successful cataract surgery in children. It is virtually inevitable if PC management is not performed at the time of primary surgery. This should be done for all children <8 years. Manual PPCCC with the help of a cystotome to make a central nick and intraocular forceps is the best recommended option [Figure 3]. A vitrector-assisted PPCCC can also be done in some cases but the margins may not be as able to resist peripheral extension of a tear
- Anterior vitrectomy may be before or after IOL placement. A shallow fast vitrectomy is preferred.[35] A limbal approach through two side ports is preferred. The purpose is just to remove the anterior vitreous within and just under PCC made as it acts as a scaffold and helps lens epithelial cell migration
- Insertion of intraocular lens in the capsular bag is best to achieve centration and optimal vision through the optic. This is carried out under cover of viscoelastic to expand the bag. Some surgeons insert an IOL first before performing the AV while some insert after. If the IOL is inserted before, retract the iris and decenter IOL slightly to get at the PC and perform the PPCCC and AV. Optic capture can also be carried out for added protection against VAO, with the optic delicately pushed through a snug PPC while the haptics remain in the capsular bag. If PPC is too large, it may decapture later. The most suitable IOL for use in young children are single piece hydrophobic foldable lens with a square edge design particularly among children <5–7 years old. It fits nicely into their small capsular bag and can fit through a very small incision making recovery very fast. It also has the lowest rates of inducing VAO. However, children older than 7 years can have a hydrophilic foldable IOL inserted. A three-piece foldable can also be used but there are higher rates of inadvertent sulcus fixation, and the haptics can kink very easily. However, single piece designs are not suitable for sulcus fixation and should be avoided as it can excite a difficult to control inflammatory response. However, foldable lens can be expensive and its use needs to be learned properly and therefore the previously used, still popular, cheap, easily available PMMA lenses can still be used after enlarging the wound to accommodate the rigid optic and it can also be placed in the sulcus if required. It is better to insert a rigid IOL properly than to insert a foldable lens wrongly. AC IOLs are not recommended in children and should be avoided even when there is no PC support due to an "accident" during surgery. A secondary IOL can however be inserted later in the sulcus or on the remnants of the now fibrotic PC rather than inserting an AC IOL
- Close all scleral tunnels/incisions with buried sutures as these children tend to rub the eyes. In addition, these wounds are less likely to self-seal and may fishmouth in children due to the elastic nature of the sclera. A "sterile" air bubble (made sterile by aspirating plain air into an empty syringe through sterile drapes or through a sterile air filter) is injected into the anterior chamber and then the main incision and side ports are "hydrated" then 10-0 nylon sutures are inserted and buried. This allows the child to open the eyes the next day for proper examination as there is no foreign body sensation.

Children <6 months or with smaller eyes should be left without an IOL. If there is no facility to perform a vitrectomy, it is appropriate to leave a child under 4 years aphakic only and do not insert an IOL till age 4 years at least or refer elsewhere to a pediatric ophthalmologist for surgery. Rates of resurgery are higher in children who have an IOL inserted in them primarily.[31]

The posterior capsular rim in these patients should be left to opacify for future support of the IOL when inserting it secondarily. Otherwise, IOL may be implanted secondarily in the sulcus.

Routinely give 0.3 ml of depot steroid and 20 mg genticin injection subconjunctivally and wipe the eye meticulously with saline.

If surgery lasts longer than 45 min, inject antibiotics intracamerally and give intravenous antibiotics also as prophylaxis against endophthalmitis.[36]

It is advised to operate the second eye within 2 weeks.

METHODS OF VISUAL REHABILITATION

1. Glasses: Aphakic glasses are not satisfactory for rehabilitation because of induced magnification, field constriction, and "jack in the box" prismatic effect they often have aside of poor compliance.[37] However, sometimes it may be the only option even in unilateral aphakia if other options are either not available or it is not yet time. It can easily scratch however and requires frequent change as the child grows
2. Contact lenses: These offer a distinct advantage such as full fields and stereopsis, but there may also be higher rates of infection, loss, costs, and difficulty with compliance in addition to poor fit[38]
3. IOL: This is the gold standard.[37,38] No IOL however should be implanted in the following cases:
   - Microphthalmos
   - Microcornea <10.5 mm
   - Axial length <17.5 mm.

CALCULATION OF INTRAOCULAR LENS POWER

SRK II formula is advocated by some to be best for children.[39,40] However, a recent report by the Infant Aphakia Treatment Study (IATS) recommends the Holladay 1 and SRK-T formula for calculation of IOL power in children.[27]

Emphasis should be on accurate axial length and K readings as for every 1 mm change in axial length measurements, there is a 3.5D change in IOL power which can cause disastrous results in visual rehabilitation.[10]

SPECIAL PROBLEMS WITH CHILDREN

1. IOL selection has remained a significant challenge[41-43]
2. Timing of insertion. Most surgeons avoid inserting an IOL in <6 months of age while some only use corneal diameter and judge whether or not to insert. In other words, the child may be 1 year old but if the horizontal corneal diameter is still <10.5 mm, then no IOL should be inserted. Some [surgeons] however wait till the child can cooperate for [accurate] biometry before inserting an IOL to reduce the rates of resurgery associated with early implantation.[31] This is somewhere in the region of 4–5 years of age. This may be expedient in countries where there is no insurance to cover repeated surgeries and where the surgeon is considered incompetent when he/she operates on an eye more than once
3. Final refractive goal: Difficult to define unlike adults.

OTHER CONSIDERATIONS

Increased tissue reactivity commonly found in children sometimes leads to severe postoperative inflammation. To minimize this, intensive topical steroids tapered over 6 weeks can be used having given an injection subconjunctivally of depot steroids at time of surgery. In addition, topical antibiotics 4 times a day can be given for 2 weeks. Avoid reoperating soon after initial surgery in the same eye also as it may excite very severe inflammation with membrane formation. Sometimes literally forming on the table and may be so severe as to occlude the pupil in some instances. Atropine ointment can be used for 4 weeks to prevent posterior synechiae formation.

FORMULAS USED TO ADJUST INTRAOCULAR LENS POWER IN KIDS

Dahan's method

- Less than 6 months of age = 35% reduction in IOL power calculated
- 6 months to 1 year = 30%
- 1–2 years = 25% reduction
- 2–5 years = 20% reduction
- 5–7 years = 10% reduction.[44]

Rule of 7 by Enyedi

Seven – age in years.[45] i.e., subtract the number of years the child is from the number 7, whatever value obtained is subtracted from the IOL power calculated. For example, a 3-year-old for whom biometry has given a value of 25D IOL power to be used should rather have an IOL power of (7 − 3 = 4. Subtract 4 from 25) 21D. This should be used as the presumed adult power and the remaining error (which should be +4D in this case) corrected with glasses till the eye has stopped growing.

Management of the posterior capsule

Preoperative

Good slit lamp examination is done to be Sure that their PC and zonules are intact. Any problem detected helps with planning surgery.

Intraoperative

Some surgeons prefer to use an AC maintainer instead of bimanual irrigation/aspiration (I/A) to avoid inadvertent rupture with the probes. In case of rupture of the posterior capsule, have ready other options of IOL models.

Make a good PPCCC (3.5–4 mm) and AV. Carry out fast vitrectomy (setting at least 600 cuts/min) to minimize hydration of the vitreous.

Postoperative

Suppress inflammation with intensive topical steroids as this excites the formation of membranes. Ensure a slit lamp examination is performed every visit looking and visual axis. Any discovery of an opacification should make a membranectomy imperative to avoid amblyopia.

Postoperative care of the patient

Keep the eye padded with the protection of a catella shield (which has a protective function by transferring any inadvertent blow to the operated eye to the forehead, temple, or nasal bridge) till the next day.

Always see the patient 1st day postoperative or ask a competent assistant to do so if you cannot at that time.

Advise the following and ensure the caregiver understands:

Topical antibiotics ×4 times daily for 2 weeks.

Topical cycloplegics for 1 month.

Topical steroids given frequently daily on a weekly taper according to inflammatory response over 6 weeks.

There is no added advantage by extending it unless there is a severe inflammatory response.

Ensure all patients are reviewed after 1 week following surgery.

Then plan after 2 weeks for examinations under anesthesia (EUA) and suture removal (side ports first, then main scleral wound later on subsequent EUAs). This minimizes inflammation. However, care must be taken to avoid inadvertent introduction of infection into the eye through the suture tracks, so it should be done under cover of povidone iodine. Avoid pulling the external parts of the suture through the tracks into the eye while pulling it out after cutting it. Children have developed endophthalmitis just from suture removal, so be careful.

Encourage meticulous ocular hygiene.

Postoperative amblyopia therapy should be meticulous and started early particularly in uniocular cataract after surgery. Amblyopia therapy is best carried out after adequate visual rehabilitation is instituted generally within 2 weeks of surgery.

Complications of cataract surgery

Amblyopia[46]

The normal eye should be occluded for up to 4–6 h daily for the formative years of the child to achieve and maintain stereopsis and binocular vision.

Strabismus[46]

This is frequent especially if amblyopia is neglected.

Visual axis opacification

Glaucoma:[47,48] Incidence varies from 3% to 32% after pediatric cataract surgery.[28] It can occur from pupil block or peripheral anterior synechiae if detected early after surgery or it may be open angle type if seen late after surgery. Hence, these children need to be followed up for life with regular IOP measurements.

Pupil capture: This occurs when a portion of the optic passes anterior to the iris. Inserting the IOL in the capsular bag reduces the likelihood of this event.

Others include the following:
Dislocated IOL (more common in can opener anterior capsulorhexis and vitrectorhexis)

Retinal detachment (RD) (especially when over exuberant anterior vitrectomy is performed or there is excessive hydration of the vitreous).

This is usually a late complication and may occur in 1–1.5% of cases.[28]

RD is more common in myopia and when repeated surgery is carried out on the same eye.[28]

Corneal opacification (when there is excessive instrumentation with poor respect for the endothelium or inadequate use of viscoelastics).

Infections (both surface and intraocular) can occur (especially when there is coexisting nasolacrimal duct obstruction), when poor personal hygiene is not taken into account or basic rules of antibacterial prophylaxis are not followed. Clean the eye 5% povidone iodine (scrub the lid margins) and use plastic drapes for eye surgery if available that stick down the lashes away from the site of surgery.

What to look out for on each follow-up visit

Visual behavior, fixation preferences/VA assessment before EUA is carried out.

Check ocular alignment using modified Krimsky or alternating prism cover test depending on the age.

Complete ophthalmic check-corneal clarity, IOL position, and central media clarity. Sometimes for the smaller children, EUAs are required due to lack of cooperation. It is best to book all cases once a week and have an anesthetist on standby to administer anesthesia. However, this may not be practical due to theater usage costs and so chloral hydrate 50–100 mg/kg (maximum dose 1000 g daily) or diazepam suppositories can be used to sedate the children.

Refraction every visit: This should be done before any other procedure to ensure enough clarity to see the reflexes properly. Take into account your working distance.

Loose sutures: This should be handled as discussed above.

IOP check with applanation tonometry.[48]

Give glasses after EUA at 3 weeks postoperative. Give near add if of school age for bifocal glasses or add the near (between + 2.00 D and + 3.00 D depending on the age) to the distance to make single vision glasses or contact lens in preschool children.

Start aggressive amblyopia therapy by patching.

EUAs every 4–6 months. Change glasses as required.

Insert a secondary IOL when at least 2 years. Maximum 4–5 years of age. This may be possible to insert in the posterior chamber or the sulcus depending on what happened in the previous surgery. Attempt to remove the "donut" of calcified cortex in between the edges of the anterior and PC remnants at the edges of the capsulotomies, otherwise the secondary IOL may not properly center.

Recent trends

1. "Bag in the lens" approach to surgery: It prevents the migration of residual lens epithelial cells to cause VAO by entrapping the AC and PC in a groove on the optic of the special lens. It requires extreme dexterity[49]
2. Use of hydrophilic acrylic IOLs: It is best for those older than 7 years[50]
3. Pediatric toric IOLs: This can be used to correct preexisting astigmatism[51]
4. Trans scleral sutured IOL lenses: In case there is no capsular support, these can be used.[24,52]

CONCLUSION

The gold standard technique of pediatric cataract surgery for best outcome is to perform manual anterior and posterior capsulorhexis in addition to using hydrophobic foldable single piece IOL through a small 3 mm sutured limbal incision. It is best to have a good strategy to treat ensuing inflammation during the postoperative period. However, predicting axial growth and the refractive change that accompanies it remains one of the major challenges for long-term care of children after cataract surgery due to the rapid growth that occurs in the first 10 years of life and invariably requires utmost care for it not to affect the eventual visual outcome.

It is recommended that adequate and close follow-up by ophthalmologists trained in pediatric eye management be encouraged and where there is difficulty, the services of social workers and ophthalmic counselors where they exist may be employed.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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